Title: The Surgical Treatment of Congenital Aortic Stenosis

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The Surgical Treatment of Congenital Aortic Stenosis

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Introduction

Congenital aortic stenosis (AS) is usually classified into three categories: supravalvular, valvular and subvalvular. Some of the problems in the surgical treatment of congenital AS, especially in the valvular and subvalvular types, are: determination of optimal age for operation, indication for various techniques, consideration for aortic commissurotomy, anticipation of sudden cardiac arrest and safety of myocardial protection, and accurate knowledge concerning etiology or long-term follow-up.

Currently, the most common technique performed for congenital AS is aortic valve replacement (AVR) instead of commissurotomy because in open commissurotomy there is a possibility that aortic regurgitation and/or aortic re-stenosis may develop, and thus reoperation may be necessary. Commissurotomy for AS with a narrow annulus is not advisable.

Recently, in congenital AS with a narrow annulus, the following methods are usually performed: aortoventriculoplasty, MANOUGUJIAN’s method and apico-aortic bypass. However, these methods need safe myocardial protection.

Problems arising in our 29 cases with congenital AS will be described herein.

Key words: Congenital Aortic Stenosis, Supravalvular Aortic Stenosis, Valvular Aortic Stenosis, Subvalvular Aortic Stenosis, Konno’s Operation.

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Materials and Methods

1) Patients

Twenty-nine patients (22 males and 7 females) with congenital AS have been operated upon since 1966 (Tables I–VI). The age range at operation was from 11 months to 43 years (mean age 14.7 years, Fig. 1).

2) Classification of AS

These cases are classified into 3 groups: 3 cases of supravalvular AS, 20 cases of valvular AS and 6 cases of subvalvular AS.

In the 3 cases of supraval!cular AS, all were male with a mean age of 5.3 years (Table I). The supravalvular AS were all of a localized type and associated with Williams elfin facies syndrome. In these cases, serum calcium values were within normal ranges at the time of operation.

In 20 cases of valvular AS, 16 patients were male and 4 were female (Tables II–IV). The mean age was 16.8 years. This group included a case in which a tumor was found on the right coronary cusp of the aortic valve (Case 23). In these patients, valvular AS was associated with the following heart diseases: single coronary artery (1), mitral regurgitation (MR) and coarctation of the aorta (1), patent ductus arteriosus (PDA) and single coronary artery (1), and mitral stenosis (MS) (2). The MS in these two cases was thought not to be congenital; the patients' ages were 23 and 41 years.

Subvalvular AS was seen in 6 patients, 3 males and 3 females, with a mean age of 12.3 years (Tables V, VI). Among the 6 cases, there was 1 case of solitary idiopathic hypertrophic subaortic stenosis (IHSS) (Case 27), 1 case of IHSS-tetralogy of Fallot (T/F) in which the T/F was corrected 9 years prior to the operation of IHSS (Case 29), and 1 case of IHSS-atrial septal defect (ASD) secundum type (II)-MR in which the ASD (II) was closed 5 months prior to the operation of IHSS and MR (Case 28). In addition, Case 29 had a membranous discrete type of subvalvular AS.

3) Surgical Procedures

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age</th>
<th>Signs</th>
<th>Symptoms</th>
<th>Chest X-P</th>
<th>ECG SV1 + RV5 (Strain pattern)</th>
<th>OP date</th>
<th>Surgical Procedures</th>
<th>Duration of A. clamping</th>
<th>Myocardial Protection</th>
<th>Ao-LV Pressure gradient (mmHg)</th>
<th>IABP</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>4 m</td>
<td>none</td>
<td></td>
<td>0.49</td>
<td>2.7 mV (—)</td>
<td>1974</td>
<td>Patch</td>
<td>5 min</td>
<td>E-CP(20C)</td>
<td>6 6 - 10</td>
<td>—</td>
<td>survived</td>
</tr>
<tr>
<td>2</td>
<td>8 m</td>
<td>none</td>
<td></td>
<td>0.49</td>
<td>2.4 mV (—)</td>
<td>1975</td>
<td>Patch</td>
<td>25 min</td>
<td>Hypothermia (23.6C)</td>
<td>20 - 5</td>
<td>—</td>
<td>survived</td>
</tr>
<tr>
<td>3</td>
<td>4 m</td>
<td>none</td>
<td></td>
<td>0.47</td>
<td>3.2 mV (—)</td>
<td>1978</td>
<td>Extended Aortoplasty</td>
<td>69 min</td>
<td>Cardioplegia ice slush (&lt;15C)</td>
<td>84 - 0</td>
<td>performed</td>
<td>survived</td>
</tr>
</tbody>
</table>

* McGoon's method  ** Doty's method

The supravalvular AS were all of a localized type and associated with Williams elfin facies syndrome.
All the supravalvular AS were treated with a Dacron patch graft, as performed by McGoon and D'Alton.

Surgical procedures for valvular AS were as follows: commissurotomy (12) (Fig. 2), aortic valve replacement (AVR) (4), aortoventriculoplasty (3), and Manouguian's method (1) (Tables 1 and 2).

Table II. Patients with Valvular AS Treated by Open Commissurotomy (n=12)

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (yr)</th>
<th>Signs/Symptoms</th>
<th>Chest X-P CTR</th>
<th>ECG SV1+RV5 (Systolic pattern)</th>
<th>OP. date</th>
<th>Associated Cardiac Disease</th>
<th>Changes in MVO before and after commissurotomy</th>
<th>Duration of Acclamping</th>
<th>Myocardial Protection</th>
<th>Ap-LV Pressure (mmHg)</th>
<th>IABP</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>4</td>
<td>16 m</td>
<td>LHF</td>
<td>0.61</td>
<td>4.8 mV +</td>
<td>1966</td>
<td>7−12mm</td>
<td>35 min</td>
<td>?</td>
<td>?</td>
<td>Died of LHF</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>18 m</td>
<td>Pulmonary</td>
<td>0.60</td>
<td>9.6 +</td>
<td>1966</td>
<td>AR (2/4)</td>
<td>36 min Ice slush CP (20min)</td>
<td>?</td>
<td>?</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>21 m</td>
<td>Dizziness</td>
<td>0.50</td>
<td>7.2 −</td>
<td>1967</td>
<td>AR (1/4) dilated 7 mm</td>
<td>10 min CP (27min)</td>
<td>12−10</td>
<td>12−10</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>21 m</td>
<td>Pericardial</td>
<td>0.45</td>
<td>4.1 (−)</td>
<td>1968</td>
<td>12−20mm</td>
<td>10 min CP (50min)</td>
<td>70−30</td>
<td>70−30</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>1 m</td>
<td>Dyspnea</td>
<td>0.69</td>
<td>7.7 +</td>
<td>1972</td>
<td>Congenital MR Coarctation of aorta 7.5−12mm</td>
<td>−</td>
<td>54−15</td>
<td>Died of Hepatitis</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>13 m</td>
<td>None</td>
<td>0.44</td>
<td>4.8 +</td>
<td>1972</td>
<td>7−15mm</td>
<td>22 min</td>
<td>12−10</td>
<td>12−10</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>13 m</td>
<td>Chest oppression</td>
<td>0.42</td>
<td>5.8 +</td>
<td>1972</td>
<td>dilated 7 mm</td>
<td>23 min</td>
<td>90−10</td>
<td>90−10</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>6 m</td>
<td>None</td>
<td>0.56</td>
<td>7.2 (−)</td>
<td>1975</td>
<td>20−22mm</td>
<td>18 min</td>
<td>30−30</td>
<td>30−30</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>11 m</td>
<td>None</td>
<td>0.47</td>
<td>4.8 +</td>
<td>1976</td>
<td>7−14mm</td>
<td>25 min</td>
<td>10−15</td>
<td>10−15</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>13</td>
<td>41 m</td>
<td>Orthopnea</td>
<td>0.51</td>
<td>2.2 (−)</td>
<td>1976</td>
<td>Rheumatic MS dilated 3 mm</td>
<td>−</td>
<td>35−5</td>
<td>35−5</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>14</td>
<td>12 m</td>
<td>Dyspnea</td>
<td>0.51</td>
<td>4.9 +</td>
<td>1977</td>
<td>dilated 6 mm</td>
<td>18 min Hypothermia (2SC)</td>
<td>15−60</td>
<td>15−60</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>15</td>
<td>3 m</td>
<td>Easy Fatigue</td>
<td>0.58</td>
<td>3.7 (−)</td>
<td>1977</td>
<td>Single coronary Artery</td>
<td>16.5 min Hypothermia (2SC)</td>
<td>15−40</td>
<td>15−40</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

MVO: Mitral Valve Orifice
Table III. Patients with Valvular AS Treated by Aortic Valve Replacement (n=4)

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age</th>
<th>Sex</th>
<th>Sign.</th>
<th>Symptoms</th>
<th>ECG OP</th>
<th>Valve orifice</th>
<th>Duration of Ao clamping</th>
<th>Myocardial Protection</th>
<th>IABP</th>
<th>Ao-LV Pressure Gradient (mmHg)</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>16</td>
<td>8 m</td>
<td>Syncope</td>
<td>0.55</td>
<td>8.7 (+)</td>
<td>1973</td>
<td>5 mm (ID) (B-S 17A)</td>
<td>114min</td>
<td>CP (120min)</td>
<td>-</td>
<td>90→10</td>
<td>died (Stone heart)</td>
</tr>
<tr>
<td>17</td>
<td>23 f</td>
<td>Syncope, Dyspnea, Dizziness</td>
<td>0.49</td>
<td>2.5 (−)</td>
<td>1974</td>
<td>8 mm (ID) (S-E 8A)</td>
<td>16min</td>
<td>CP (70min)</td>
<td>-</td>
<td>95→10</td>
<td>died (Stone heart)</td>
</tr>
<tr>
<td>18</td>
<td>43 m</td>
<td>Dyspnea, Chest oppression, Palpitation</td>
<td>0.51</td>
<td>7.8 (+)</td>
<td>1976</td>
<td>0.3cm² (B-S 21ABP)</td>
<td>64.5min</td>
<td>CP (50min)</td>
<td>-</td>
<td>70→5</td>
<td></td>
</tr>
<tr>
<td>19</td>
<td>38 m</td>
<td>Edema, Dyspnea</td>
<td>0.68</td>
<td>6.0 (−)</td>
<td>1977</td>
<td>8 mm (ID) (L-K 18A)</td>
<td>12min</td>
<td>CP (100min)</td>
<td>+</td>
<td>70→5</td>
<td></td>
</tr>
</tbody>
</table>

CP: Coronary Perfusion

II-IV. Aortoventriculoplasty was performed in 3 cases with valvular AS (Table IV).

Cases 20 and 21 survived the operation, but Case 22, having PDA and a single coronary artery, died of low cardiac output syndrome (LOS). Retrograde cold blood cardioplegia was performed on Case 22, but the contraction of the right ventricle (RV) was too poor to decrease central venous pressure. The duration of aortic cross-clamping was 108 minutes. In Case 21,

Table IV. Patients with Valvular AS Treated by AVR Combined with Annular Enlargement (n=4)

<table>
<thead>
<tr>
<th>Patient (age, sex)</th>
<th>20 (13yo. m)</th>
<th>21 (12yo. f)</th>
<th>22 (6yo. m)</th>
<th>23 (11yo. m)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diagnosis</td>
<td>Valvular AS R¹(1)*</td>
<td>Valvular AS R¹(2)*</td>
<td>Valvular AS R¹(1)*, PDA Single coronary artery</td>
<td>Valvular AS R¹(1) due to dysplasia, MR, PH</td>
</tr>
<tr>
<td>Symptoms</td>
<td>none</td>
<td>none</td>
<td>dizziness</td>
<td>LHF</td>
</tr>
<tr>
<td>CTR</td>
<td>0.51</td>
<td>0.58</td>
<td>0.58</td>
<td>0.74</td>
</tr>
<tr>
<td>SV, +RV (mm²)</td>
<td>7.5</td>
<td>8.5</td>
<td>5.7</td>
<td>8.1</td>
</tr>
<tr>
<td>Strain pattern</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
</tbody>
</table>

Cardiac catheterization

<table>
<thead>
<tr>
<th></th>
<th>preop</th>
<th>postop</th>
<th>preop</th>
<th>postop</th>
<th>preop</th>
<th>postop</th>
<th>preop</th>
<th>postop</th>
<th>preop</th>
<th>postop</th>
</tr>
</thead>
<tbody>
<tr>
<td>LV 198/0,18</td>
<td></td>
<td></td>
<td>240/0,14</td>
<td></td>
<td>240/0,20</td>
<td></td>
<td>175/0,20</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ao 100/70</td>
<td>100/60 (80)</td>
<td></td>
<td>85/65</td>
<td>124/88</td>
<td>110/60 (85)</td>
<td></td>
<td>80/45 (63)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>m-PA 31/15 (21)</td>
<td>30/20 (25)</td>
<td></td>
<td>22/12 (17)</td>
<td>40/20 (30)</td>
<td>36/17 (22)</td>
<td></td>
<td>70/15 (38)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>RV inf 45/5,15</td>
<td>48/8,8</td>
<td></td>
<td>32/5</td>
<td>60/2,20</td>
<td>36/8,0</td>
<td></td>
<td>70/0,10</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>


Surgical procedure

Konno’s method | Konno’s method | Konno’s method | Manouguian’s method

Prosthesis

Bjork-Shiley 23 AB | Bjork-Shiley 23 ABP | Omura-Science 21 A | Bjork-Shiley 17 ABP

Myocardial protection

Coronary perfusion (115 min) | Cardioplegia (MIK) Topical cooling | Cardioplegia (Cold Blood) Topical cooling | Cardioplegia (MIK) Topical cooling

Duration of Aortic clamping | 25min | 100min | 108min | 116min

IABP

Performed |Performed |Performed |Not performed

Postop. course

RHF, TR (3%), Residual VSD (Shunt 41.5%) | RHF, Contraction of RV: poor | RHF, Contraction of RV: poor | LHF

Results

Survived | Survived | Died (4 days postop.) | Died (16 days postop.)
moderate tricuspid regurgitation and residual ventricular septal defect (VSD) remained postoperatively. All three of these cases suffered from LOS due to right heart failure, to which intra-aortic balloon pumping (IABP) was performed postoperatively. The size of the prosthesis for Cases 20–22 were adequate (Björk-Shiley valve 21 and 23, and Omni-Science 21, respectively). Myocardial protection was respectively performed using the following techniques: coronary perfusion (115 min.), cardioplegia using MIK solution (100 min.), and cold blood cardioplegia (108 min).

In Case 23, AVR (Björk-Shiley 17 A) was performed, enlarging the aortic annulus by the

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age</th>
<th>Sex</th>
<th>Signs</th>
<th>Symptoms</th>
<th>Chest X-P</th>
<th>ECG SV₁ + RV₁s</th>
<th>OP. date</th>
<th>Type of Subvalv. AS</th>
<th>Surgical Procedure</th>
<th>Duration of Aortic clamping</th>
<th>Myocardial protection</th>
<th>Ao-LV Press.</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>24</td>
<td>5 m</td>
<td>none</td>
<td>0.51</td>
<td>7.4 (+)</td>
<td>1976</td>
<td>Discrete (Kelly I-II)</td>
<td>transaortic resection 7 → 14 mm</td>
<td>24 min</td>
<td>Hypothermia by H-L bypass (28°C)</td>
<td>130–40 survived</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>25</td>
<td>6 m</td>
<td>syncope</td>
<td>0.53</td>
<td>8.3 (−)</td>
<td>1977</td>
<td>Membranous discrete (Kelly I)</td>
<td>transaortic resection 6.5 → 13 mm</td>
<td>22 min</td>
<td>Hypothermia by H-L bypass (32°C)</td>
<td>70–32 survived</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>26</td>
<td>10 f</td>
<td>none</td>
<td>0.48</td>
<td>8.2 (+)</td>
<td>1977</td>
<td>Discrete (Kelly I)</td>
<td>transaortic resection 8 → 20 mm</td>
<td>20 min</td>
<td>Hypothermia by H-L bypass (32°C)</td>
<td>50–22 survived</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Fig. 2. Patients with Valvular AS Treated by Open Commissurotomy (n = 12)
Manouguian's method. This size of the prosthesis was the smallest one which we could obtain at that time. As MR (Grade 2–3) had been detected preoperatively, damage to the mitral valve was avoided. Cardioplegia using MIK solution was performed, with a duration of aortic cross-clamping of 116 min. However, the patient did not survive the operation due to left heart failure resulting from residual MR.

In all cases with subvalvular AS, myotomy was performed (Tables V, VI), and when subvalvular AS was associated with the other heart diseases, total repair was performed.

Results

All patients with supravalvular AS survived the operation, however, in valvular AS and subvalvular AS, 6 patients (30%) and 3 patients (50%), respectively, died; total mortality was 9 of 29 (31.0%). In valvular AS, 6 patients died of the following causes: left heart failure (2), hepatitis (1), stone heart (2) and LOS (1). All three patients with subvalvular stenosis associated with IHSS died; 2 operative death and 1 late death. In valvular AS, the surgical procedures for the cases which did not survive were: commissurotomy (2), AVR (2), aortoventriculoplasty (1) and Manouguian's method (1). Case 4 had a severe narrow annulus and slight regurgitation; he underwent an insufficient commissurotomy in 1966, but subsequently died of left heart failure. Case 8 had severe mitral regurgitation, coarctation of the aorta and slight aortic regurgitation; he underwent aortic commissurotomy and mitral annuloplasty, but died of acute hepatitis 3.5 months postoperatively. Except for these 2 cases, all patients who underwent commissurotomy survived the operation. The duration of aortic cross-clamping, in which commissurotomy was performed, was less than 40 minutes. However, AVR required aortic clamping of more than 60 minutes. Therefore, cold coronary perfusion, topical cooling (ice slush), and cardioplegia were performed for myocardial protection. The two patients who died of AVR required more than

Table VI. Patients with IHSS (n=3)

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age-</th>
<th>Signs-</th>
<th>Chest X-P</th>
<th>ECG</th>
<th>OP</th>
<th>Surgical procedures</th>
<th>Duration of Ao. clamping</th>
<th>Myocardial Protection</th>
<th>IABP</th>
<th>Ao-LV Press.</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>27</td>
<td>20</td>
<td>Syncope</td>
<td>0.65</td>
<td>7.8</td>
<td>1968</td>
<td>Myotomy through RV</td>
<td>75 min</td>
<td>CP (65min)</td>
<td></td>
<td></td>
<td>64–16</td>
</tr>
<tr>
<td>28</td>
<td>24</td>
<td>Palpation</td>
<td>0.53</td>
<td>4.2</td>
<td>1972</td>
<td>Closure of ASD (I)</td>
<td>*</td>
<td>*</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>29</td>
<td>9</td>
<td>Anoxic spell</td>
<td>0.56</td>
<td>6.5</td>
<td>1977</td>
<td>Total correction for T/F</td>
<td>*</td>
<td>*</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Results

All patients with supravalvular AS survived the operation, however, in valvular AS and subvalvular AS, 6 patients (30%) and 3 patients (50%), respectively, died; total mortality was 9 of 29 (31.0%). In valvular AS, 6 patients died of the following causes: left heart failure (2), hepatitis (1), stone heart (2) and LOS (1). All three patients with subvalvular stenosis associated with IHSS died; 2 operative death and 1 late death. In valvular AS, the surgical procedures for the cases which did not survive were: commissurotomy (2), AVR (2), aortoventriculoplasty (1) and Manouguian's method (1). Case 4 had a severe narrow annulus and slight regurgitation; he underwent an insufficient commissurotomy in 1966, but subsequently died of left heart failure. Case 8 had severe mitral regurgitation, coarctation of the aorta and slight aortic regurgitation; he underwent aortic commissurotomy and mitral annuloplasty, but died of acute hepatitis 3.5 months postoperatively. Except for these 2 cases, all patients who underwent commissurotomy survived the operation. The duration of aortic cross-clamping, in which commissurotomy was performed, was less than 40 minutes. However, AVR required aortic clamping of more than 60 minutes. Therefore, cold coronary perfusion, topical cooling (ice slush), and cardioplegia were performed for myocardial protection. The two patients who died of AVR required more than
## Table VII. Patients Who Did Not Survive the Operation (n=9)

<table>
<thead>
<tr>
<th>Patient (age, sex)</th>
<th>Diagnosis</th>
<th>Operation Procedures</th>
<th>Date</th>
<th>Cause of Death</th>
<th>Time of Death</th>
<th>Duration of Ao. clamping</th>
</tr>
</thead>
<tbody>
<tr>
<td>4 (16yo. m)</td>
<td>AS R(1') severe narrow annulus</td>
<td>Comm.</td>
<td>1966</td>
<td>LHF</td>
<td>OP. day</td>
<td>35 min</td>
</tr>
<tr>
<td>8 (1 yd. m)</td>
<td>AS Bicuspid MR Coarctation of the aorta</td>
<td>Comm. MAP</td>
<td>1972</td>
<td>Hepatitis</td>
<td>3.5 mo postop.</td>
<td>24 min</td>
</tr>
<tr>
<td>16 (8 yo. m)</td>
<td>AS LV cavity small LV wall 20mm</td>
<td>AVR (B-S 17)</td>
<td>1973</td>
<td>“Stone heart”</td>
<td>Table death</td>
<td>80 min CP 45min intermittent</td>
</tr>
<tr>
<td>18 (43yo. m)</td>
<td>AS Bicuspid Calcified</td>
<td>AVR (B-S 21)</td>
<td>1976</td>
<td>“Stone heart”</td>
<td>Table death</td>
<td>70 min CP 50min intermittent</td>
</tr>
<tr>
<td>22 (6 yo. m)</td>
<td>AS R(1') PDA Single coronary artery</td>
<td>Konno’s OP. (O-S 21) Ligation of PDA</td>
<td>1981</td>
<td>LOS</td>
<td>4 days postop.</td>
<td>116 min MIK Cardioplegia</td>
</tr>
<tr>
<td>23 (11mo. m)</td>
<td>AS R(1') Valvular dysplasia MR. PH</td>
<td>Manouguian’s OP (B-S 17)</td>
<td>1981</td>
<td>RHF</td>
<td>16 days postop.</td>
<td>75 min CP 65min</td>
</tr>
<tr>
<td>27 (20yo. f)</td>
<td>IHSS</td>
<td>Myotomy through RV</td>
<td>1968</td>
<td>LHF (?</td>
<td>5 years postop.</td>
<td>—</td>
</tr>
<tr>
<td>28 (25yo. f)</td>
<td>ASD (I)</td>
<td>Closure of ASD (I)</td>
<td>1972</td>
<td>5</td>
<td></td>
<td></td>
</tr>
<tr>
<td>29 (9 yo. m)</td>
<td>IHSS MR</td>
<td>1) Myotomy through RV 2) MVR S−E 3M.</td>
<td>1972</td>
<td>LOS</td>
<td>Op. day</td>
<td>66 min</td>
</tr>
<tr>
<td>29 (9 yo. m)</td>
<td>T/F</td>
<td>Total repair</td>
<td>1968</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>29 (9 yo. m)</td>
<td>IHSS</td>
<td>1) Transcatheter closure of discrete membrane 2) Myotomy through RV</td>
<td>1977</td>
<td>LOS</td>
<td>Table death</td>
<td>80 min CP 34min intermittent</td>
</tr>
</tbody>
</table>

Comm : Commissurotomy LHF : Left Heart Failure CP : Coronary Perfusion MAP : Mitral Annuloplasty B−S Björk-Shiley S−E Starr-Edward

120 minutes of aortic cross-clamping, with coronary perfusion of 45 minutes (Case 16) and 50 minutes (Case 18).

Concerning postoperative changes in the LV-Ao pressure gradient, supravalvular AS was superior to subvalvular AS (Fig. 3). In all 3 types of AS, a decrease was seen in the LV-Ao pressure gradient (mmHg) between the pre- and postoperative values. In supravalvular AS, it decreased from $56.6\pm26.9$ to $5.0\pm4.1$; in valvular AS, from $88.9\pm33.8$ to $17.9\pm16.5$; in subvalvular AS, from $91.5\pm32.6$ to $27.0\pm9.5$. 
Discussion

Accurate knowledge as to the etiology of congenital AS and hemodynamic change while the patient grows is an important problem in the surgical treatment of congenital AS. The incidence of congenital AS is 2.5–3% of congenital heart diseases. EL-Said and Cohen reported that severity of AS is progressive while the patient grows. However, Hurwitz reported that no hemodynamic changes could be found in several long-term studies.

As to the age distribution of congenital AS, there are no characteristic features between the ages of 1 year and 15 years. However, in patients over 20 years old, it is possible that congenital AS may be associated with acquired valvular diseases, for example rheumatic mitral stenosis or subacute bacterial endocarditis. The majority of the patients with valvular AS can survive infancy without symptoms, or, with only slight symptoms, however, there is a great possibility of sudden cardiac arrest before the occurrence of symptoms. Therefore, the presence of symptoms is of little value in the determination of operation time.

1) Supravalvular AS

Edwards classified supravalvular AS into 3 groups, namely 1) membranous, 2) hourglass type and 3) hypoplastic type. Our three cases were of the hourglass type, and were treated by
a Dacron patch, as described by McGo0n and Doty; the results were good. The hypoplastic type of supravalvular AS should be treated by apico-aortic bypass.

2) Valvular AS

There are five basic types of congenital aortic valvular stenosis: unicuspid, bicuspid, tricuspid (dome-shaped), undifferentiated, and membranous. Lillehei stated that from the surgical viewpoint, the infantile group (under 1 year of age) presents different problems from the remaining groups. The cases that require surgery in the first year of life are usually characterized by two features: the valves are almost always unicuspid and, except for endocardial fibroelastosis, associated anomalies such as aortic valvular tumor (Case 23) are rarely encountered. Histological studies revealed that the tumor was a dysplasia of the aortic valve. Usually, the operative criteria for congenital AS are: 1) LV-Ao pressure gradient more than 50 mmHg; 2) a valvular orifice area less than 0.5 cm²/BSA(M²); 3) a strain pattern by electrocardiography; and 4) recurring anginal or syncopal attack.

The key point for achieving good results with open commissurotomy is whether the aortic valve has a wide annulus and no calcium deposits. Postoperative results of open commissurotomy are not always good, because slight regurgitation and/or a small LV-Ao pressure gradient may still remain. Follow-up studies with electrocardiography have not shown good late results. However, open commissurotomy is the safest method for enlarging the aortic valvular orifice. Usually, in a severely hypertrophied left ventricle there is a limitation in the duration of aortic clamping and also a great possibility of stone heart due to insufficient myocardial protection. Except for one case (Case 4) with a severe narrow annulus, no patient died following open commissurotomy. Thus open commissurotomy can be considered a safe palliative method. Calcium deposits in the aortic valve, which frequently need AVR, are rare in childhood; but increase in frequency in adults. In our cases, 4 of 16 patients required AVR without annular enlargement. Since AVR needed a longer duration of aortic clamping (over 120 min), than open commissurotomy, its mortality (2/4) was higher than that of commissurotomy (2/12). Of the two AVR patients who died, both died of 'stone heart' on the operation table. In Case 16, the LV cavity was very small with a wide LV wall of 20 mm. In Case 18, the aortic valve was bicuspid and calcified. When a narrow annulus is present, a method which enlarges the annulus is necessary in order to achieve good results.

Recently, in congenital AS with a narrow annulus, the following methods are usually performed:

1) Nicks and Blank's method, in which the non-coronary cusp is incised extending to the mitral annulus.
2) Konno, Sohma and Rastan's method (aortoventriculoplasty)
3) Manougian's method in which the annulus is inserted at the site between the non- and left coronary cusp, extending to the anterior mitral cusp.

Rastan operated upon patients with the following lesions: 1) isolated diffuse fibromuscular subaortic stenosis, 2) diffuse subaortic stenosis plus other cardiovascular anomalies, 3) hypoplastic aortic annulus, 4) IHSS, and 5) previously implanted small aortic valvular prosthesis.
These methods achieve sufficient enlargement of the annulus. However, prosthesis and long duration of aortic clamping are necessary. Improvement of prosthesis and cardioplegic techniques has made this complex surgery easy and safe. However, the surgical damage is extensive enough to require some strong cardioplegic techniques. During the operation we commonly used cold coronary perfusion or topical cooling (ice slush) with cold cardioplegic solution (MIK solution21). In addition, IABP was used during the operation and postoperatively.

In AS, a severely hypertrophied LV wall, high LV pressure, low cardiac output, increased oxygen consumption and cardiac work may bring about myocardial damage and a relative ischemic state of endocardium due to decreased coronary blood flow5. Especially in cases associated with a single coronary artery, it may be difficult to completely perform cardioplegic techniques. Cardioplegic solution cannot be infused because, as the solution escapes from the incision of the right ventricle and interventricular septum, the infusion pressure drops resulting in a decrease of infusion flow, especially in the endocardial layer.

KIMOTO19 reported the experimental results of various surgical procedures which enlarge the aortic annulus using live mongrel dogs and extirpated porcine hearts in order to study the clinical usefulness. According to his findings, MANOUGIAN'S19 method could enlarge the aortic annulus with a rate of enlargement of 17.1±4.3% without mitral regurgitation. On the other hand, aortoventriculoplasty could enlarge the aortic annulus with a rate of enlargement of 23.6±2.5% with safety.

The youngest patient whom RASTEN23 performed successful operation was 8 years old. Thus, from experimental and clinical studies, the optimal age, in which an adult size prosthesis can be inserted with safety using aortoventriculoplasty, appears to be over 8 years. However, with a single coronary artery, there is a limitation in the selection of surgical procedures for enlarging the aortic annulus.

WADA27 stated that, when the aortic root is hypoplastic and severe calcium deposits are present in the aortic valve, apico-aortic bypass3-22 is advisable. Furthermore, he pointed out the following indications for apico-aortic bypass: 1) insufficient alleviation of aortic stenosis, 2) insufficient myotomy in subvalvular AS, 3) reoperation following AVR and/or MVR, and 4) inapplicability of surgical procedure to aortic valve due to subacute bacterial endocarditis. This method may be advisable for reoperation following AVR in infancy or childhood, as in Case 23 of aortic tumor.

Recently, with improvement of cardioplegic techniques and prosthesis, AVR combined with enlargement of annulus is performed instead of open commissurotomy. However, patients less than 8 years old should be treated by commissurotomy as a first choice of surgical procedure.

3) Subvalvular (subaortic) AS

There are two types of subvalvular AS: 1) discrete subvalvular stenosis or subvalvular fibrous ring stenosis, 2) idiopathic hypertrophic subaortic stenosis (IHSS).

In discrete subvalvular stenosis, transaortic resection of the obstructive portion is advisable. In our three cases, transaortic myotomy was performed and good results were achieved. However, there is a combined type of discrete subvalvular stenosis and hypertrophied muscular stenosis.
In IHSS, indications for surgical treatment are of outmost importance. Frank and Braunwald reported that the natural course of IHSS was variable. Therefore, accurate criteria for operative indication can not be established. However many authors agree with the following criteria: 1) severe symptoms, 2) no improvement by administration of propranolol, 3) severe stenosis at rest and increased stenosis upon exertion, and 4) remarkable increase of stenosis by administration of isoproterenol.

Resection of obstructive muscle has been performed by various approaches: 1) transaortic, 2) through LV-apex, 3) through RV, and 4) through LA. We experienced two IHSS cases associated with other heart diseases. One had T/F which was treated by total repair 9 years previously. The other had ASD(II), which was treated 6 months previously, and MR. Both cases were treated by resection of the obstructive muscle through right ventriculotomy. However surgical damage was so extensive that the patients did not survive the operation. Our case of T/F was a combined type of discrete subvalvular stenosis and IHSS. Rastan operated upon two cases of IHSS by aortoventriculoplasty, and good results were achieved in one case, but the other died of severe acidosis during perfusion.

**Summary**

In supravalvular AS, except for the hypoplastic type, the use of a Dacron patch poses no problem in surgical treatment. The hypoplastic type should be treated by apico-aortic bypass.

In valvular AS, with improvement of prosthesis and cardioplegic techniques, AVR which enlarges the aortic annulus has been performed with safety. However surgical damage is so extensive that strong myocardial protection is necessary to achieve good results. These methods have a limitation in the rate of enlargement of the aortic annulus, thus limiting the patients' age to over 8 years old. The patient less than 8 years old should be treated by commissurotomy, a safe palliative operation, as a first choice of surgical procedure until the time when radical operation can be performed.

In subvalvular AS, the membranous discrete type was treated by transaortic resection of the obstructive muscle with good results. However, other types of subvalvular AS, including IHSS, should be treated by aortoventriculoplasty. In IHSS, clear surgical indications are necessary in order to achieve good results.

**References**


先天性大動脈狭窄症の外科治療

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大動脈弁狭窄症では，低形式型を除いてパッチ拡大にて良好な成績を得た。
大動脈弁狭窄症では，弁置換術が主流となりつつあるが，交換弁術の高い安全性は再考の余地がある。
成人用サイズの人工弁置換の困難な 8 才患者では，第1選択として交換弁術か，8 才以上では今野の手術をはじめ人工弁置換術がより良策と考えられる。
大動脈弁狭窄症では，discrete 型では，経大動脈弁切除で良好な結果が得られたが，HSS では，結果は不恵で，手術適応の確立がますます必要である。